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Case Report
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Visual Loss and Proptosis as Ocular Manifestation of Extranodal Natural Killer/T-Cell Lymphoma, Nasal Type

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Abstract

Extranodal NK/T-Cell Lymphoma (ENKTCL) is extremely rare, and only a few cases have reported so far. We report the case of a 33-year-old female patient who presented with blurred vision and periorbital swelling and then proven to have ENKTCL.

Keywords: Visual loss; Proptosis; NK/T-cell lymphoma

Introduction

Extranodal NK/T-Cell Lymphoma (ENKTCL) is a rare type of Non-Hodgkin Lymphoma (NHL) and deviated from either activated NK cells or cytotoxic T cells [1]. This type of lymphoma is most common in people from Asia and Central and South America with a male predominance [2]. About 80% of cases occur in the nose, nasopharynx, oropharynx, and upper aero digestive region. Another 20% occur in non-nasal sites, including the skin, testis, gastrointestinal tract, muscle, and salivary glands [3]. The neoplasm presents with initial nonspecific nasal symptoms, nasal discharge, nasal obstruction, rhinitis, sinusitis, cellulitis, hemifacial pain, and edema. We report a case of ENKTCL, nasal type, suffered from unilateral visual loss and proptosis as an ocular manifestation.

Case Presentation

A-33-year-old female noted blurred vision and periorbital swelling in the left eye. She also suffered from nasal obstruction intermittently. Her past medical histories included diabetes mellitus under medical control and bilateral chronic rhino sinusitis status post bilateral pansinusectomy. She denied any trauma history. On examination, visual acuity was 20/20 in the right eye and no light perception in the left eye. Left eye movement was limited and afferent pupillary defect was noted. Ocular fundi were normal. Nasal fiberscopy showed bilateral nasal mucous and bilateral posterior choanal stenosis with synechia. Laboratory examinations showed white blood cell count and percentage of neutrophils elevated moderately (white blood cells: $8.3x10^3$/uL; neutrophils: 79.3%). The Epstein-Barr virus titer showed positive. Brain Magnetic Resonance Imaging (MRI) (Figure 1) scan revealed enhancing and infiltrating lesions involved left nasal cavity, left ethmoid sinus, orbital apex and extra-conal region, left cavernous sinus, and meningeal of left anterior temporal base. Para nasal sinus biopsy by functional endoscopic sinus surgery was performed. Histopathological examination of the excised tissue showed atypical lymphocytic infiltrate with enlarged nuclei, irregular nuclear border and angiocentric pattern with focal coagulative necrosis. Immunophenotypically the tumor cells were positive for CD56, CD8, Ki-67, negative for cytokeratin, CD3 and CD20. Subsequent bone marrow aspiration also shows infiltrate of atypical lymphocytes and positive for CD 56. In-situ hybridization study revealed tumor cells harboring EBV-RNA. The histological and immunohistochemical analysis were compatible with the diagnosis of ENKTCL, nasal type, complicated with retrobullar compressive optic neuropathy .The whole body PET scan showed the increase F18 Floro Deoxy Glucose (FDG) metabolism at left maxillary region, nasal region, bilateral cervical lymph nodes, liver, spleen, axial and appendicular skeletons. A definite diagnosis of ENKTCL, stage IV was made and then the patient received chemotherapy. Unfortunately, due to neutropenia and multiple organ failure, she expired one month later.

Discussion

ENKTCL is a rare and aggressive malignancy. Most patients with ENKTCL were relatively young and most commonly affects age of between 40 and 50 years, with a male predominance [2]. ENKTCL accounts for 7% to 10% of NHL cases in Asian and Latin American countries, but only 1% of Caucasian cases [4,5]. The pathogenesis is not understood completely but Epstein - Barr Virus (EBV) is considered as sine qua in this type of lymphoma [6]. The lymphoma is locally invasive.

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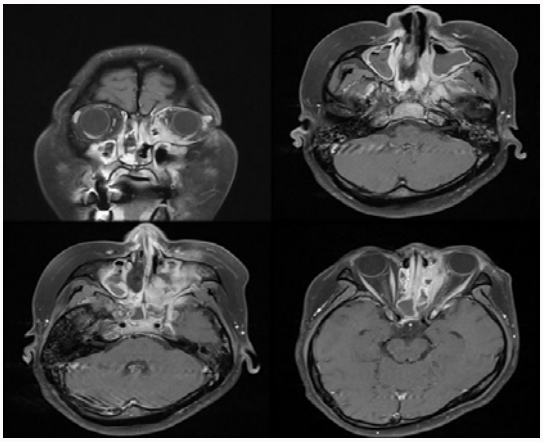

Figure 1: Brain MRI revealed enhancing and infiltrating lesions involved left nasal cavity, left ethmoid sinus, orbital apex and extra-conal region, left cavernous sinus, and meningeal of left anterior temporal base on T1-weighted image.

Because of angio-destruction, the lymphoma destroys midline facial structures and involves hard palate, orbit, salivary glands, and para nasal sinuses [7]. Symptoms of nasal type ENKTCL can include nasal discharge, nasal obstruction and other nonspecific sinonasal symptoms. However, the presence of visual loss and periorbital swelling has not yet been reported. ENKTL is both chemo sensitive and radio sensitive. However, the prognosis is poor if ENKTCL spread to multiorgan involvement, including spleen, liver, and bone marrow [8]. The WHO classification defines as aggressive NK-cell leukaemia [1]. Clinical course is relentlessly downhill, with survival time often measured only in weeks to months.

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References

1. Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, et al. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissue (4th ed.). IARC Press. 2008;276-7.
2. Aozasa K, Zaki MA. Epidemiology and pathogenesis of nasal NK/T-cell lymphoma: a mini-review. Scientific World Journal. 2011;11:422-8.
3. Tse E, Kwong YL. The diagnosis and management of NK/T-cell lymphomas. Journal of Hematology & Oncology. 2017;10:85.
4. Quintanilla-Martinez L, Kremer M, Keller G, Nathrath M, Gamboa-Dominguez A, Meneses A, et al. p53 mutations in nasal natural killer/T-cell lymphoma from Mexico: Association with large cell morphology and advanced disease. Am J Pathol. 2001;159(6):2095-105.
5. Au WY, Ma SY, Chim CS, Choy C, Loong F, Lie AK, et al. Clinicopathologic features and treatment outcome of mature T-cell and natural killer-cell lymphomas diagnosed according to the World Health Organization classification scheme: A single center experience of 10 years. Ann Oncol. 2005;16(2):206-14.
6. Gru AA, Haverkos BH, Freud AG, Hastings J, Nowacki NB, Barrionuevo C, et al. The Epstein-Barr Virus (EBV) in T Cell and NK Cell Lymphomas: Time for a Reassessment. Curr Hematol Malig Rep. 2015;10(4):456-67.
7. Metgud RS, Doshi JJ, Gaurkhede S, Dongre R, Karle R. Extranodal NK/T-cell lymphoma, nasal type (angiocentric T-cell lymphoma): A review about the terminology. J Oral Maxillofac Pathol. 2011;15(1): 96-100.
8. Kwong YL. The diagnosis and management of extranodal NK/T cell lymphoma, nasal type and aggressive NK cell leukemia. J Clin Exp Hematop. 2011;51(1):21-8.